

Neuroimaging of a Paediatric Ophthalmoplegic Migraine

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CLINICAL DESCRIPTION

A 10-year-old male presented with a seven-day history of right hemicranial headache, one episode of emesis, complaints of visual blurring, and intermittent double vision since five days. He had no systemic symptoms or prior medical illnesses. No prior history of similar episodes, trauma, or a diurnal variation was reported. Upon examination he was afebrile with a normal sensorium. No meningeal signs were noted. Fundus examination was normal. Right third nerve paresis with ptosis, “down and out” position of the right eyeball [Figure 1a] with sparing of right lateral rectus and superior oblique muscles, and a poorly reacting right pupil was noted. Other cranial nerves were uninvolved. No long tract signs, hemiparesis, or other focal neurological deficits were noted. Peripheral nervous system examination was unremarkable. Sepsis screen, vasculitic work up, creatinine phosphokinase, thyroid function test, routine haematological, and biochemical parameters were unremarkable. Study of cerebrospinal fluid was declined by the family. A magnetic resonance imaging (MRI) was performed. Focal nodular thickening and contrast enhancement of the right 3rd cranial nerve medial to the cerebral peduncle near the exit zone was seen [Figure 1b and 1c]. Rest of the cisternal, cavernous and orbital parts of the right 3rd nerve were

normal. No other brain parenchymal abnormalities were seen. A diagnosis of ophthalmoplegic migraine (OM) was made. The child was initiated on oral steroid. There was a clinical improvement and complete resolution of right 3rd nerve paresis was seen on the fifth day of starting steroid.

DISCUSSION

OM is a misnomer with a controversial etiology and is now considered to be a demyelinating cranial neuropathy. The international classification of headache disorders—IHCD III Beta classifies it as recurrent painful ophthalmoplegic neuropathy (RPON) with features of ipsilateral headache, subsequent cranial neuropathies, specific MRI features, and steroid responsiveness.^[1,2] The etiology is either a schwannoma or recurrent demyelination of the affected cranial nerve.^[3]

Recurrent episodes, transient, and reversible focal contrast enhancement of third nerve, complete resolution of imaging findings and clinical remission with response to steroid support an underlying demyelinating pathology.

The clinical features are atypical for migraine with long latent period up to 14 days before the onset of ophthalmoplegia and prolonged headache up to 1 week or more.^[2] The third cranial nerve is most often involved followed by 6th and 4th cranial nerves. Notable differences in adult OM/RPON include single episode with no recurrence, common 6th nerve involvement, worsening of migraine during or 24 h prior to developing the ophthalmoplegia, and often a normal neuroimaging.^[4]

Congenital palsy remains the commonest cause of 3rd nerve paresis in children (43%) followed by trauma (20%) and focal inflammation (17%). Other notable causes were myasthenia gravis and OM/RPON. Although less common in children (7%), aneurysm, especially involving the posterior communicating artery, is one of the most important life threatening cause of 3rd nerve paresis that should not be missed. Other important differentials that should be ruled out include 3rd nerve schwannoma, granulomatous disorders, like sarcoidosis, and infiltrative disorders, like lymphoma.^[5]

Focal thickening and enhancement of the affected cranial nerve may be demonstrated on a MRI (reference). The root exit zone of the third nerve is typically affected, as in this case. Both thickening and enhancement may persist, but enhancement usually resolves over time.^[3,6]

Recovery may be spontaneous but is hastened by a steroid treatment.

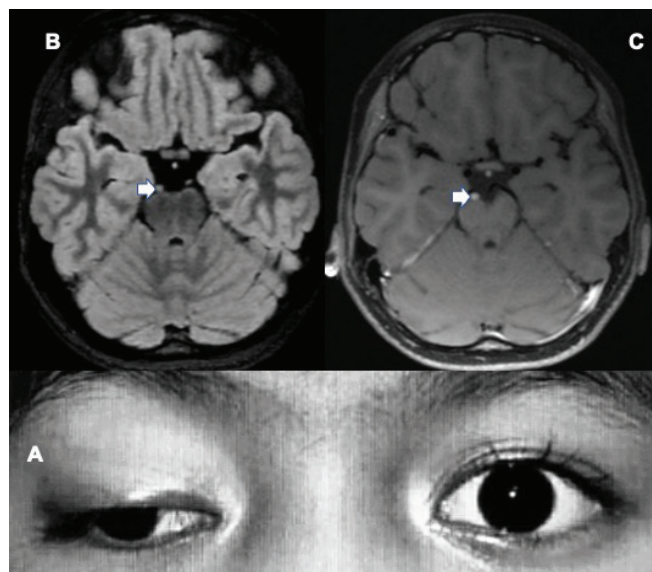


Figure 1: (a) Right eye ptosis with a “down and out” position of the eyeball typical of a third nerve palsy. The left eye is normal. (b) FLAIR axial MRI image showing focal nodular thickening of the right 3rd cranial nerve (bold arrow) at the exit zone. (c) Post contrast axial MRI image shows focal contrast enhancement of the right 3rd nerve (bold arrow).

The diagnosis of RPON/OM is generally made clinically with MRI serving as an important adjunct to diagnosis. Follow up to look for recurrence is warranted as recurrent episodes are common in children.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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